<https://doi.org/10.47660/CBR.2020.17514>

LIVEDOID VASCULOPATHY A THERAPEUTIC CHALLENGE: CASE REPORT

Ricardo Amaro Noletto Araujo^{1,2,*}, Sany Tomomi de Almeida Rocha Arita², Samy Simantob Fridman², Raquel de Nardo Claudino², Mirela Bianchezi Fernandes², Alisson Lisboa Cordeiro², Giovanna Garcia Stabile²

1.Hospital Santa Marcelina, São Paulo (SP), Brazil. 2.Faculdade Santa Marcelina, São Paulo (SP), Brazil.

*Corresponding author: ricardoreumatologista@gmail.com

BACKGROUND

Livedoid vasculopathy (LV) manifests with chronic and recurrent skin lesions, characterized by ulcerations and edema of the lower limbs. It manifests with painful erythematous or purpuric macules and/or papules, associated with itching on legs and ankles, in 3/4 months, bringing about a white atrophy in a star shape¹. Female predominance (3:1) and average age of 32 years (between 15 and 50 years)². The study aims to report the case of LV that obtained adequate therapeutic response with the administration of cyclophosphamide³.

CASE REPORT

Male patient, 25 years old, machine maintenance technician, without comorbidities. Approximately 3 years ago, purple spots started to appear on his legs and ankles, pruritic and slightly painful. The episode evolved with worsening, appearance of painful ulcers, about 2 months before the first consultation in medical service, in this period in use of vasodilators, possibly treating as chronic venous disease. Physical examination presented ulcers in legs with areas of healing affecting 1/3 of the skin, with itching and pain. In the complementary examinations, patient with normal inflammatory tests with negative antinuclear factor (ANF) and antineutrophil cytoplasmic antibody (ANCA), skin biopsy with stasis dermatitis and fibrin thrombi in vessels of the dermis with discrete perivascular inflammatory infiltration. The use of oral anticoagulant warfarin was initiated, and INR 2–3 was obtained. The patient evolved in 2 months with worsening of the lesions, appearance of necrosis and larger area of ulceration (Fig. 2). Due to refractivity, pulse therapy cyclophosphamide 1.0 g for 6 months was associated. Cyclophosphamide therapy improved ulcerations and pain control in the first 2 pulse therapies (Fig. 3). The patient maintained the medicine warfarin; however, the average INR of 1.3 indicates low adherence (Fig. 4).

Figure 1. Period in use of vasodilators, possibly treating as chronic venous disease.

Figure 2. In 2 months with worsening of the lesions, appearance of necrosis and larger area of ulceration.

Figure 3. Pulse therapy cyclophosphamide 1.0 g for 6 months was associated. Cyclophosphamide therapy improved ulcerations and pain control in the first pulse therapies.

Figure 4. Patient maintained the medicine warfarin.

CONCLUSION

Livedoid vasculopathy is a disease whose pathophysiology is not well elucidated. Its clinical manifestation can assume a primary character, not related to comorbidities, or secondary, which can be associated to thrombophilic episodes. Clinical investigation must ascertain the history of inflammatory diseases and the state of hypercoagulability. It is believed that the pathophysiological mechanism is due to a vaso-occlusive process caused by thrombosis of dermal vessels. Inflammatory manifestations are initially marked by neutrophilic and late lymphocytic content. Some treatments have been pointed out in case reports such as the of anti-TNF-alpha 2 or series of cases such as anticoagulants, steroids, antiplatelet agents, among others. Cyclophosphamide is associated with favorable outcome and improvement of the lesions in the short term. Nonadherence of warfarin with possible recurrence or nonresponse factor is noted. This report reinforces the need for further studies to treat this pathology.

REFERENCES

1. Provenza JR, Pedri LE, Provenza GM. Livedoid vasculopathy. *Rev Bras Reumatol.* 2016;6(6):554–6. <https://doi.org/10.1016/j.rbr.2015.09.011>
2. Zanini M, Wulkan C, Bertino D, Ito L. Livedoid vasculopathy or vasculitis? *An Bras Dermatol.* 2003;78(6):755–7. <https://doi.org/10.1590/S0365-05962003000600012>
3. Miciel R, Alavi A. Treatment for livedoid vasculopathy: a systematic review. *JAMA Dermatol.* 2017;154(2):193–202. <https://doi.org/10.1001/jamadermatol.2017.4374>